Cell Junctions in the Specialized Conduction System of the Heart.

Journal: Cell Commun Adhes

Publication Year: 2014

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PubMed link: 24738884

Funding Grants: Molecular Mechanisms Underlying Human Cardiac Cell Junction Maturation and Disease Using Human iPSC

Public Summary:
Anchoring cell junctions are integral in maintaining electro-mechanical coupling of working muscle cells; however, their role in cardiac muscle cells of the conduction system (CCS) remains less clear. Recent studies in genetic mouse models and humans highlight the appearance of these cell junctions alongside electrical (gap) junctions in the CCS and also show that defects in these structures and their components are associated with conduction impairments in the CCS. Here we outline current evidence supporting an integral relationship between anchoring and gap junctions in the CCS. Specifically we focus on (1) molecular and ultrastructural evidence for cell-cell junctions in specialized cardiac muscle cells of the CCS, (2) genetic mouse models specifically targeting cell-cell junction components in the heart which exhibit CCS conduction defects and (3) human clinical studies from patients with cell-cell junction-based diseases that exhibit CCS electrophysiological defects.

Scientific Abstract:
Anchoring cell junctions are integral in maintaining electro-mechanical coupling of ventricular working cardiomyocytes; however, their role in cardiomyocytes of the cardiac conduction system (CCS) remains less clear. Recent studies in genetic mouse models and humans highlight the appearance of these cell junctions alongside gap junctions in the CCS and also show that defects in these structures and their components are associated with conduction impairments in the CCS. Here we outline current evidence supporting an integral relationship between anchoring and gap junctions in the CCS. Specifically we focus on (1) molecular and ultrastructural evidence for cell-cell junctions in specialized cardiomyocytes of the CCS, (2) genetic mouse models specifically targeting cell-cell junction components in the heart which exhibit CCS conduction defects and (3) human clinical studies from patients with cell-cell junction-based diseases that exhibit CCS electrophysiological defects.